

## CASE REPORT

## Pediatrics

# Superior mesenteric artery syndrome: A review of the literature

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**Funding and support:** By *JACEP Open* policy,  
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as per ICMJE conflict of interest guidelines (see  
[www.icmje.org](http://www.icmje.org)). The authors have stated that  
no such relationships exist.

JE and NL contributed equally to this article.

**Abstract**

Decreased intraabdominal fat can lead to intraabdominal compressive syndromes, such as superior mesenteric artery (SMA) syndrome. This phenomenon is rare but should be considered in a patient with recent rapid weight loss and acute gastrointestinal complaints. A delay in diagnosis and treatment can lead to severe complications, such as a gastric rupture. We report a case of SMA syndrome in a teenage male with recent intentional weight loss and intractable emesis, and the possible associations of SMA syndrome and Nutcracker syndrome.

**KEYWORDS**

Bowel obstruction, Nutcracker syndrome, SMA syndrome, Superior mesenteric artery

## 1 | CASE

A 15-year-old male presented to the emergency department (ED) with abdominal pain and vomiting for 3 days. He described the pain as severe (8 out of 10), sharp, and mostly epigastric. He reported that the pain transiently improved after vomiting, which had occurred 4 times in the last 24 hours (non-bloody and non-bilious). He also reported decreased frequency of bowel movements (1 in the last 7 days).

The patient was seen at an outside ED on day 1 of illness and was diagnosed with constipation. He was prescribed docusate, ondansetron, and magnesium citrate. He then presented to our ED on day 2 of the illness for continued abdominal pain, vomiting, and no bowel movements. An abdominal radiograph was obtained, showing a mild volume of colonic stool, a non-obstructive bowel gas pattern, and a severely distended stomach filled with debris. The patient reported improvement in his symptoms after receiving an enema and having a moderate-sized bowel movement. He was discharged again with the diagnosis of constipation.

He returned to the ED on day 3 of symptoms for continued pain and vomiting, despite using ondansetron and docusate as prescribed. Addi-

tional review of systems revealed that the patient had intentionally lost  $\approx 60$  pounds (27.2 Kg) in 6 months by "cutting out junk food."

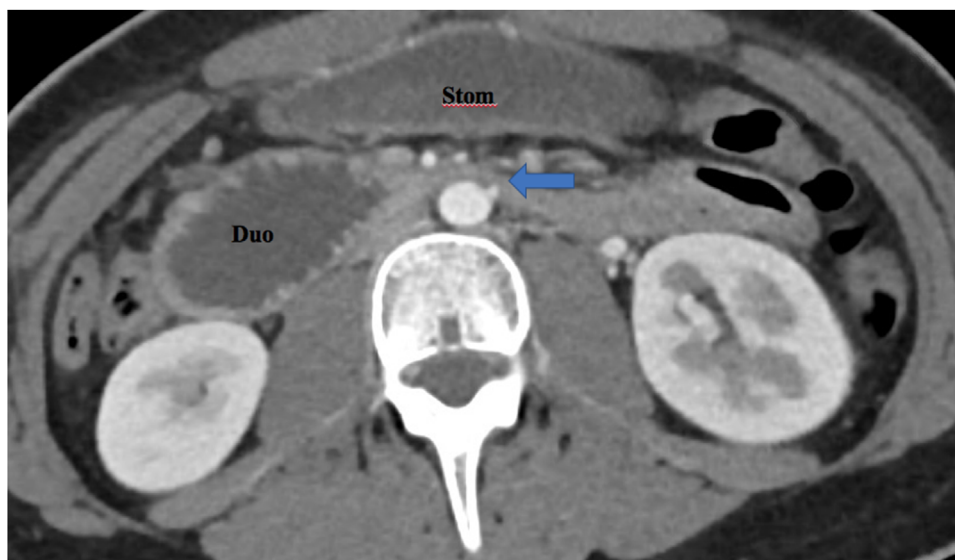
His vital signs were as follows: temperature of 37.9°C, heart rate of 116 beats per minute, respiratory rate of 25 breaths per minute, blood pressure of 133/83 mm Hg, and oxygen saturation of 99% on room air. His weight was 64.9 kg (71st percentile on the Centers for Disease Control and Prevention [CDC] growth chart), with a body mass index (BMI) of 19 kg/m<sup>2</sup>. Physical examination revealed an alert and cooperative male with dry mucous membranes, tachycardia but no murmurs, and normal respiratory effort. He was noted to be tall (96th percentile on the CDC growth chart) with evidence of joint hypermobility: when grasping his opposite wrist, his thumb overlaps with his fifth finger (a positive wrist sign). Abdominal examination revealed tenderness to palpation at the epigastric area. Normal bowel sounds were present. He had no tenderness over McBurney's point, the right upper quadrant, or the costovertebral margins. Genital examination showed normal Tanner Stage 4 male genitalia, with no evidence of testicular swelling or tenderness.

Laboratory evaluation showed a hypochloremic metabolic alkalosis, with a chloride of 97 mEq/L (normal: 98–107 mEq/L) and a bicarbonate of 34 mEq/L (normal: 22–30 mEq/L). Blood urea nitrogen was mildly elevated at 22 mg/dL (normal: 8–21 mg/dL), but creatinine was normal.

Supervising Editor: Benjamin T. Kerrey, MD, MS.

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**FIGURE 1** An axial view of an abdominal computed tomography (CT) shows a severely distended stomach (Stom) anteriorly. A distended third part of the duodenum (Duo) is seen before it passes under the superior mesenteric artery (arrow)

Total bilirubin was elevated at 2.3 mg/dL (normal < 1.3 mg/dL), with direct bilirubin of 0.0 mg/dL (0.0 mmol/L). Sodium, potassium, glucose, calcium, phosphorus, liver enzymes, lipase, and C-reactive protein were within normal limits. A urinalysis was unremarkable.

A computed tomography (CT) scan of the abdomen was obtained and revealed prominent distension of the stomach and proximal duodenum, with abrupt transition to a decompressed caliber in the third segment of the duodenum adjacent to the superior mesenteric artery (SMA) origin. The angle between the proximal SMA and the abdominal aorta was narrow, at 16°, and there was dilation of the left renal and gonadal veins with left varicocele. These findings were consistent with SMA syndrome and Nutcracker syndrome.

## 2 | DISCUSSION

Constipation is a common cause of abdominal pain in the pediatric ED and should typically be a diagnosis made from history and physical alone. However, it also can be a source of misdiagnosis, especially when clinicians anchor on the diagnosis. A study by Freedman et al<sup>1</sup> showed patients were more likely to be misdiagnosed with constipation if an abdominal radiograph was obtained, highlighting a possible undervaluation of history and physical examination and overvaluation of an abdominal radiograph. Study after study has been done to discourage the use of abdominal radiograph in the diagnosis of constipation.<sup>2-6</sup>

Alternative diagnoses, including an obstructive process, should be considered in patients with severe abdominal pain, persistent emesis, or bilious emesis. A prolonged absence of stooling, especially without straining or other specific evidence of constipation, and rapid weight loss should be especially concerning for SMA syndrome.

In SMA syndrome (also known as Wilkie's syndrome), rapid weight loss decreases the aortomesenteric distance because of a reduction of important intraabdominal fat, specifically the duodenal fat pad. The normal aortomesenteric angle ranges from 28° to 65° and the nor-

mal aortomesenteric distance ranges from 10 to 34 mm.<sup>7</sup> In SMA syndrome, the SMA leaves the abdominal aorta at an abnormally acute angle, typically < 22–25°, and runs closer to the aorta than normal, typically < 2–8 mm depending on the patient's BMI.<sup>8</sup> No number is strictly diagnostic and imaging must be interpreted in light of the clinical history.

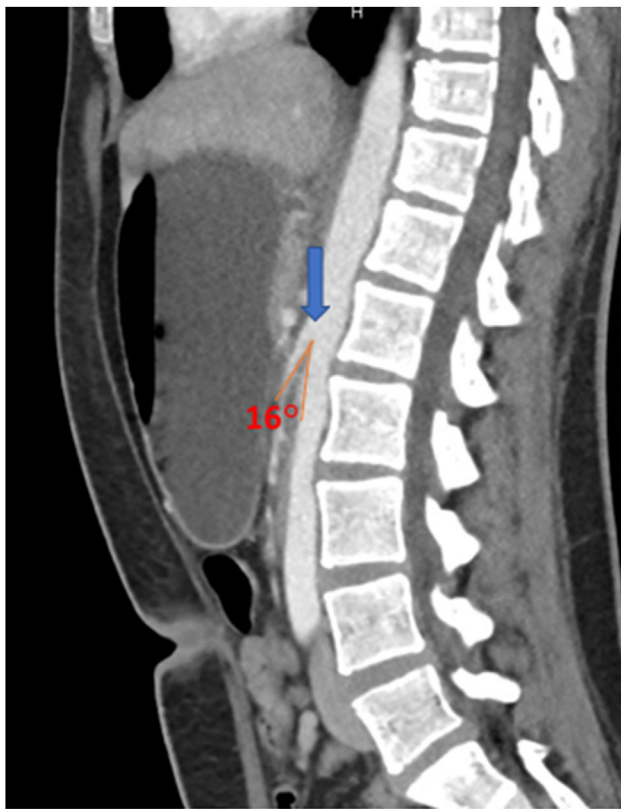
The hyperacute angle that the SMA makes with the aorta compresses the third segment of the duodenum, leading to the classic presenting findings of severe epigastric abdominal pain,<sup>9</sup> gastric distension, early satiety, and vomiting.<sup>10</sup> The literature discusses cases of weight loss from anorexia nervosa being associated with SMA syndrome and even the likelihood of delayed diagnosis of these entities in males because of a common gender bias of eating disorders.<sup>11,12</sup> In pediatrics, the reported co-occurrence of SMA syndrome and Nutcracker syndrome is relatively rare,<sup>13-15</sup> but it is not unexpected that they would occur together. Both conditions are due to compression of structures at similar anatomical levels within the abdomen.

Nutcracker syndrome is due to compression of the left renal vein because of narrowing between the SMA and the abdominal aorta, which can eventually lead to left flank pain, hematuria, proteinuria, and/or a left varicocele.<sup>8,16</sup> Rarely, Nutcracker syndrome also can be related to a normal variant retroaortic left renal vein that gets compressed by the aorta anteriorly and the vertebral body posteriorly, but this version is much less common than the classic anterior form.<sup>17,18</sup> Nutcracker syndrome can be associated with other anatomical pathologies, such as gut malrotation or, as in our patient's case, SMA syndrome.<sup>8</sup>

This possible association of SMA and Nutcracker syndrome with Ehlers-Danlos syndrome (EDS) has also been described,<sup>19</sup> so this triad should be considered in other similarly presenting tall, overly flexible patients.

Diagnosis of both SMA and Nutcracker syndrome is made based on history and abdominal imaging, especially CT abdominal imaging. On axial CT images, the vascular compression of the duodenum, with a

**FIGURE 2** An axial view of an abdominal computed tomography (CT) shows a severely distended stomach (Stom) can be seen anteriorly. An enlarged left renal vein (star) can be seen and is consistent with Nutcracker syndrome



**FIGURE 3** A sagittal view of an abdominal computed tomography (CT) shows a narrowed angle of  $\approx 16^\circ$  (arrow) between the superior mesenteric artery and the abdominal aorta. The severely distended fluid-filled stomach can be seen anteriorly in this image



**FIGURE 4** A sagittal view of an abdominal computed tomography (CT) shows a normal angle (arrow) of the superior mesenteric artery coming off the abdominal aorta

proximally distended duodenum, will be seen in SMA syndrome<sup>13</sup> (Figure 1). In Nutcracker syndrome, the “beak sign” of a narrowed left renal vein may be observed (Figure 2). The approximation of the angle of the SMA coming off the abdominal aorta can best be made using sagittal cuts on the abdominal CT (Figures 3 and 4). Secondary findings, such as a varicocele in Nutcracker syndrome or distension of the stom-

ach in SMA syndrome, can also be seen. Upper gastrointestinal fluoroscopy in the supine position can also diagnose an obstructed duodenum, whereas doppler ultrasonography in a somewhat upright position may visualize left renal vein obstruction.<sup>8</sup>

The mainstay of treatment in SMA and Nutcracker syndromes is refeeding and weight gain. Surgical intervention is indicated only if refeeding and weight gain fail to relieve the renal vascular congestion and abdominal pain symptoms.<sup>9</sup> Patients typically do well with

enteral tube feeds until small frequent meals are eventually tolerated by mouth. Ultimately, the goal is to regain lost intraabdominal fat and prevent rare but severe complications, including gastric perforation in untreated SMA syndrome or renal vein thrombosis in untreated Nutcracker syndrome. Electrolyte abnormalities should resolve with simple rehydration, especially hypochloremic metabolic alkalosis.

The patient was admitted to the hospital and a nasogastric tube was placed for gastric decompression. Subsequent nasojejunal tube (NJT) placement for enteral feeds led to a 6.7 kg weight gain during his inpatient stay, and was discharged home with continued NJT feeds. Follow-up ultrasound after his initial weight gain showed complete resolution of the renal vein compression. The patient was eventually diagnosed with anorexia nervosa and is continuing care with outpatient eating disorder specialists. At the time of this report, he also was scheduled for a genetic evaluation for EDS.

In emergency medicine, hesitancy to perform CT imaging because of concern for radiation exposure should be balanced with the need for CT to diagnose rare but relatively serious disease processes. SMA should be considered in patients with recent rapid weight loss and persistent vomiting and/or abdominal pain. CT imaging is the preferred approach to diagnosis. Management is typically conservative, with most patients responding to refeeding and weight gain.

#### ACKNOWLEDGMENTS

We would like to thank Dr. Joseph England for his contributions in the editing process and his radiology expertise.

#### CONFLICT OF INTEREST

The authors declare no conflict of interest.

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**How to cite this article:** England J, Li N. Superior mesenteric artery syndrome: A review of the literature. *JACEP Open*. 2021;2:e12454. <https://doi.org/10.1002/emp2.12454>